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ABSTRACT

A case study focuses on the characteristics and physical management of a 15-year-old with Prader-Willi Syndrome, a birth defect associated with hypotonia, insatiable appetite, hypogonadism, central nervous system dysfunction, and abnormal growth and development . A literature review addresses studies dealing with behavior modification of obesity and continuous eating, nutrition counseling, and physical activity intervention. Interventions in the case study included those programed by trained personnel in the following service areas: adapted physical education, behavior modification, nutrition, nursing, dormitory living, social work, education, and speech and language. Data are reported for behavior, weight, and adjustment to physical activity. Analysis of data from the S's developmental and medical histories, continuous evaluations, previous staffing summaries, motor proficiency tests, exercise metabolism tests, progress reports and staff and parent survey forms revealed that the S made considerable progress in reaching her program goals. She was found to demonstrate most of the characteristics classic to the Prader-Willi population. Recommendations were made for programming. (CL)

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PRADER-WILLI DISEASE: A CASE STUDY

by

WILLIAM R. FORBUS, III

B.S., Georgia College, 1982

An Applied Project in Physical Education

at the University of Georgia

in Partial Fulfillment

of the

Requirements for the Degree

MASTER OF EDUCATION

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PRADER-WILLI DISEASE: A CASE STUDY

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Approved:

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December 8, 1983
Date

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CHAPTER I

INTRODUCTION

Due to recent legislation, regular physical educators have found more handicapped students integrated into their classes. These handicapping conditions are coupled with such secondary types of conditions as obesity. This has compounded the physical educator's problem in appropriate service delivery. Correlated with the influx of these handicapped children in the regular physical education class has come the advent of the health related physical fitness movement, which has affected physical educators by paying increasing attention to such factors as muscular strength/endurance, cardiovascular endurance, and body composition; including body fatness.

Of these handicapped children, there is an incidence of obesity found in some children, that has come to be known as a syndrome. This incidence, termed Prader-Willi Syndrome, now includes some children who were formerly characterized as obese. These students have insatiable appetites and eat continuously. If not restrained, they become more obese and less active, thus heightening the self-defeating cycle.

Because of the many parameters encompassing Prader-Willi Syndrome, a cooperative effort appears necessary between a number of professionals in education and related therapeutic services. Following an interdisciplinary approach in treating these students, an adequate and successful program may be implemented.

Physical exercise without caloric intake control has limited results and diet alone cannot alleviate the problem. A cooperative effort between all professionals involved (i.e., the treatment team) is essential in providing adequate service to meet programming needs. Specifically, interventions such as physical activity, nutrition, counseling, behavior modification, leisure activity, parent and community training will help ensure successful service delivery.

Parent and community training should be an integral part of programming so the intervention will be a continuous 24-hour process. Knowledge and skills learned in the school setting should carry over into the home and community settings. By training parents and related service personnel in the community, a continuity in the total lifestyle of the Prader-Willi student may be found.

Appropriate programs are needed in physical education/adapted physical education for Prader-Willi students. Exercise programs that reflect objectives of decreasing body fat, improving mobility, improving the child's self-concept and self-image will enhance the total educational service delivery in meeting program goals (i.e., permanent and sustained weight loss and increased physical activity). Therefore, a case study with a review of current literature and detailed developmental history would aid in constructing appropriate programs for these persons. Not only will the subject benefit from this study, but some broad generalizations on treatment may arise.

Statement of the Problem

As the identification of persons with Prader-Willi Syndrome rises, there is a need for studies in the areas of physical activity, nutrition

counseling, and behavior modification. The purpose of this case study is to investigate and chronolog the status and experiences of a diagnosed Prader-Willi child, and to determine how the child compares with current research and literature.

Limitations

This case study is limited to the child's immediate environment. It is also limited to the resources found in historical documents and medical/educational records that are specific to the child.

Basic Assumptions

For the purpose of this study, the investigator will assume that all records and data collected on the child are true and accurate.

Definition of Terms

Hyperphagia- excessive overeating

Hypogonadism- incomplete or abnormal sexual development

Hypotonia- reduction or lack of muscle tone

Jejunoileal bypass surgery- bypass surgery of the second section of the small intestine in order to enhance loss of weight

Obesity- an increase in body weight beyond skeletal or physical requirements, as the result of excessive accumulation of fat

Polyphagia- an abnormal or excessive desire for food

Prader-Willi Syndrome- a birth defect characterized by hypotonia, insatiable appetite, hypogonadism, central nervous system dysfunction, and abnormal growth and development

CHAPTER II

REVIEW OF LITERATURE

Introduction

A syndrome is a group of signs or symptoms that collectively indicate a disease or disorder. Prader-Willi Syndrome was originally described in 1956 by doctors Andrea Prader, Alexia Labhart, and Heinrich Willi (Bray, Dahms, Swerdloff, Fiser, Atkinson, & Carrell, 1983; Schultze, 1979; Wannarachue, Ruvalcaba, & Kelley, 1975). It is a rare birth defect manifested by hypotonia, mental deficiency, hypogonadism, shortness of stature, marked obesity (Bray et al., 1983; Butler, Weaver, & Meany, 1982; Wannarachue et al., 1975), as well as other complications. Incidence is not clearly known because of the undiagnosed cases; however, figures range from "over 100" (Thompson, Kodluboy, & Heston, 1980, p. 588) to "fewer than 500" (Schultze, 1979, p. 2594-A). The syndrome may be more prevalent than previously thought, with recent estimates of incidence between 1 in 5,000 and 1 in 10,000 (Holm, Sulzbacher, & Pipes, 1981).

Since there is presently no known cure for Prader-Willi Syndrome, treatment is directed at the associated difficulties using an interdisciplinary approach. One report suggested using a three-prong school-based program for all obese children (Brownell & Kaye, 1982). Since obesity is a major characteristic and problem associated with Prader-Willi Syndrome, indications are that a focus should be on these three area: behavior modification, nutrition counseling, and physical activity (Brownell & Kaye, 1982).

Prader-Willi Syndrome

The etiology of Prader-Willi Syndrome cannot be attributed to any single factor or group of factors. Specific features of the syndrome such as hypogonadism have been studied in an attempt to find their etiology. Earlier studies attributed hypogonadism to hypothalamic or pituitary dysfunction; however, anumber of studies have pointed toward D chromosome abberations/translocations/deletions as a possible cause of Prader-Willi Syndrome. Data indicate, however, that hypogonadism in Prader-Willi Syndrome is not necessarily hypothalamic/pituitary in origin and that D chromosome abberations/translocations/deletions are not sufficient to explain that cause of the syndrome (Wu, Hasen, & Warburton, 1981).

Rare reports of multiple affected siblings or of parental sanguinity have raised the possibility of the syndrome being autosomal recessive inheritance (Bonuccelli, Statten, Levitt, Levin, & Pyeritz, 1982). Guanti (1980) brought up another possibility: "an autosomal dominant mutation with lack of familial recurrence caused by hypogonadism" (p. 423).

There have been several reports documenting a translocation in the D or G series of chromosomes or alternatively the presence of a small amount of extra-chromosomal material (Buhler, Rossier, Bordis, Vulliet, & Stalder, 1963; Cohen & Gorlin, 1969; Dunn, Ford, Averspberg, & Miller, 1961).

Most cases reported are sporadic in families, which suggests multifactorial causation or of exposure to an unknown teratogen (Bonuccelli et al., 1982). Four patients with translocations at the q band on chromosome 15 were reported by Emberger, Rodiere, Astruc, and Brunel

(1977) and Fracarro, Zuffardi, Buhler, and Jurik (1977). Bray et al. (1983) found chromosomal patterns that showed a deletion or translocation at chromosome 15 in 3 of 12 patients tested. Another report revealed a 13/15 chromosome abberation in a 14-year old male with Prader-Willi Syndrome (Wu et al., 1981). The patient's mother and five of six siblings had a balanced Robertsonian translocation that was a 13/15 abberation, and only the patient had Prader-Willi Syndrome. Therefore, the association of translocation in chromosome 15 with Prader-Willi Syndrome is not straightforward.

Other D chromosome cases have been reported (Fugita, Sakamoto, & Hamamoto, 1980; Kozma, Scribanu, Baumiller, Cochran, & Kenessey, 1980; Kucerova, Strakova, & Polivkova, 1979; Ridler, Garrod, & Berg, 1971) and it has been suggested that deletion of a specific band from the chromosome 15 short arm might be a specific etiology of Prader-Willi Syndrome (Fleischnick, Cone, Greer, & Wood, 1979).

Another study showed that a simple translocation is not the etiology of the syndrome. In a case study of a female Prader-Willi Syndrome patient with a 14/15 translocation, Smith and Noel (1980) found three normal family members with the same chromosomal configuration confirmed by G and C banding.

Cases have been reported in all races (Bray et al., 1983; Endo, Tasaka, Matsuura, & Matsuda, 1976); however, in the literature reviewed describing specific cases of Prader-Willi Syndrome, only two of the individuals were identified as blacks (Butler et al., 1982).

Behavior Modification

Nearly 100 percent of the Prader-Willi Syndrome cases reported developed polyphagia, characterized by continuous eating and an insatiable appetite. Temper tantrums, outbursts of anger, moodiness, stealing food, eating garbage, and erratic, unpredictable behavior have been noted often triggered by thwarting food demands (Otto, Sulzbacher, & Worthington-Roberts, 1982; Prader/Willi Syndrome Parents & Friends, 1977; Thompson et al., 1980).

Crnic, Sulzbacher, Snow, and Holm (1980) did research involving 32 diagnosed Prader-Willi children. Behavior modification plans were designed for each family, specific to the child's needs. Consultants, including social workers and endocrinologists, were made available to families on clinic days. The focus of treatment was on weight maintenance and management of food-related behavior problems. Results indicated that an early-intervention program appears to improve the children's prognoses.

Holm and Pipes (1976) and Pipes and Holm (1973) reported an approach at the University of Washington which employed behavior modification and involved precise control of caloric intake and the utilization of a protein sparing diet. This diet was later studied by Bistrain, Blackburn, and Stanbury (1977) to investigate further use of very low carbohydrate levels in dietary management.

A 1979 study reported treatment of four young adults with Prader-Willi Syndrome in a closed clinical research unit (Marshall, Elder, O'Basky, & Liberman). Patients were taught to subdivide meal portions of a caloric reduction diet and consume the smaller of the portions.

Eating unapproved foods between meals resulted in loss of the next meal. Additional privileges were contingent upon weight loss and frequency of meal violations. Success of the program appeared to relate to the close monitoring of the research unit. Research has indicated that Prader-Willi patients have been fairly unresponsive to all but very intensive treatments involving close monitoring.

Tiller (1983) established an immediate reward behavior management plan for a Prader-Willi patient utilizing a daily checklist. If the patient was successful on one section of the checklist, she received immediate positive reinforcement. Reinforcements consisted of activities that were favorable and appropriate to the student. Results indicated improved compliance, eating and exercise habits, and weight loss.

Thompson et al. (1980) noted that social isolation combined with aversion therapy and point loss therapy suppressed weight increases in a Prader-Willi patient. On the discontinuation of either therapeutic approach, rapid weight increase (75 pounds over 2 months) was noted. Results indicated that excessive overeating was not contingent upon social reinforcing consequences.

Another study of Prader-Willi patients showed that self-monitoring and contingency contracting produced a 26 percent loss of body weight in two individuals treated as outpatients (Altman, Bondy, & Hirsch, 1978). With another Prader-Willi patient, Thompson et al. (1980) concluded that close supervision and a point loss contingency program for inappropriate eating was the most effective procedure in obtaining sizable weight reductions.

Nutrition Counseling

Most notable from a nutritional point of view is the obesity that develops without intervention strategies. Hyperphagia, being a major characteristic of Prader-Willi Syndrome, is not the sole contributing factor towards obesity. Coplin, Hine, and Gormican (1976) indicated that Prader-Willi children must be restricted to fewer calories than healthy, non-affected children to maintain their weight in appropriate growth channels; therefore, further reduction in caloric intake must be instituted to reduce weight.

Twelve Prader-Willi summer campers were used in a research project to correlate activity levels and weight loss (Nardella, Sulzbacher, & Worthington-Roberts, 1983). A 1,000 caloric diet consisting of protein (23%), fat (23%), and carbohydrates (54%) was utilized in conjunction with a planned physical activity program. Weight loss may have been attributed directly to the increase in exercise or physical activity; however, results seemed to indicate a combination of diet and physical activity.

Bahling (1979), in a case study of two Prader-Willi patients, stated that the success of training appetite control was doubtful due to the physiological involvement of the syndrome. She suggested scheduled meals, snacks, and low calorie foods as a more appropriate intervention for compulsive eating. However, a report by Touquet, Ward, and Clark (1983) indicated that a steady progressive weight loss may be achieved in Prader-Willi patients whom no change in eating habits was expected. This was accomplished by jejunoileal bypass surgery. One Prader-Willi patient lost 62 kilograms during the year following surgery. Zellweger and Schneider (1968), in the analysis of one group of Prader-Willi patients, noted a

reduction in the degree of overweightness to a value of 170 percent which approximated the group not having undergone surgical procedures. The use of surgical intervention should be used when weight cannot be controlled by other methods (Bray et al., 1983).

Hoefnagel, Costello, and Hatoum (1967) stated that quality and choice of food was clearly secondary to quantity in their Prader-Willi patients. Another study reported that for this population "quality of food seemed to be less important than quantity" (Zellweger & Schneider, 1968, p. 592). Caldwell and Taylor (1983), however, found that Prader-Willi Syndrome individuals did have a definite, consistent food preference; and further, the degree of choice was related to the level of the cognitive ability of the individual. They also found that their patients chose lesser amounts of preferred food over greater amounts of non-preferred food (Caldwell & Taylor, 1983).

Dietary therapy in older Prader-Willi patients has had limited success in controlling hyperphagia. One recent study seems to indicate that early diagnosis and institution of dietary therapy and nutrition counseling ameliorate later complications (Bonuccelli et al., 1982).

Holm and Pipes (1976) found that 11 of their 14 Prader-Willi patients lost weight on calorie-restricted diets. Crnic et al. (1980) stated that maintaining a lower body weight would improve the progress as measured by development quotient; and, therefore, individuals maintained at a lower body weight may have higher developmental quotients than obese individuals.

In a correlative study, Coplin et al. (1976) gave their patients a choice of four diets. Mothers tended to reduce carbohydrates even further than instructed by the dietician. Bistrain et al. (1977) found success

in using a very low calorie diet consisting primarily of protein and a reduction of carbohydrates.

The focus of treatment of obesity in the syndrome should be on weight control and management of food-related behavior problems (Crnic et al., 1980). Long-term weight losses are based on the changes of eating habits combined with physical activity (Brownell & Kaye, 1982). Nutrition education or counseling is an integral part of a number of programs in treating obesity in non-Prader-Willi adolescents and adults as well as Prader-Willi individuals (Coates & Thoresen, 1978; Epstein, Wing, Koeske, Andrasik, & Ossip, 1981; Seltzer & Mayer, 1970).

Physical Activity

The most common approaches in the treatment of obesity in Prader-Willi Syndrome have been nutrition counseling, pharmacotherapy, therapeutic starvation, and exercise instruction. Coates and Thoresen (1978) found that meaningful weight losses were rare and that long-term maintenance of these losses was even rarer. Using a combined approach of nutrition counseling, behavior modification, and physical activity, Brownell and Kaye (1982) compared two groups of obese children. Their program was based on the concept that changes in eating and exercise habits were the key to long-term weight losses. Results indicated that 95.2 percent of the program children lost weight, compared to 21.4 percent of the control children. "The mean changes in percentage overweight were a decrease of 15.4 percent for the program children and an increase of 2.8 percent for the control children (Brownell & Kaye, 1982, p. 280).

In 1970, Seltzer and Mayer reported modest weight loss in a five-month program (3 classes per week) in elementary and junior high school

students using a combination of exercise, nutrition education, and psychological support. More recently, a ten-week program of behavior modification, exercise, and nutrition counseling by Botvin, Cantlon, Carter, and Williams (1979) produced significant weight losses in non-Prader-Willi adolescents.

Bahling (1979) reported in her two case studies that neither patient enjoyed formal exercise. By documenting participation in motor skill activities, it was found that most Prader-Willi children avoided active physical exercise in their play (Carman, 1981).

In a study of 40 patients, Bray et al. (1983) examined pulmonary function tests in patients with Prader-Willi Syndrome as compared with normal weight and moderately obese men and women. Results indicated a marked reduction of vital capacity, expiratory volume, inspiratory capacity, functional reserve capacity, and total lung capacity in Prader-Willi patients. Residual volume was noted to be the same. "Single breath oxygen (SBO_2) was the same as normal whereas in other obese subjects it was depressed" (Bray et al., 1983, pp. 64-65).

Bullen, Reed, and Mayer (1964), Oschi (1973), and Stunkard and Pestka (1962) found the differences in activity levels of obese and non-obese normal children. The obese children were found to be significantly less active than the non-obese children.

Carman (1981) noted a perpetuating cycle in Prader-Willi patients. "Low muscle tone and poor coordination result in inactivity and a low energy output; inactivity encourages a reduction in muscle strength and coordination. To complicate this cycle, Prader-Willi Syndrome children want to eat anytime food is available, and they need less calories than

non-affected children" (Carman, 1981, p. 499). He further stated that the cycle can be broken by the introduction and encouragement of daily physical activity (Carman, 1981).

Nardella et al. (1983) introduced an intervention strategy at a summer camp for Prader-Willi children. Attempts were made to interrupt the aforementioned cycle through a recreation program that emphasized physical exercise. Activity levels were measured with actometers and pedometers of the campers and compared with a control group. In a two-week period, all campers lost 0 - 36 kilograms. More weight was lost the first week than the second week. "In general, the Prader-Willi Syndrome campers who lost the most weight were not the most physically active" (Nardella et al., 1983, p. 502). No significant correlation was found between weight change and actometer readings; however, a significant correlation was found between weight change and pedometer readings. They also suggested that self-motivation and positive reinforcement were powerful influences on activity levels in Prader-Willi students (Nardella et al., 1983).

CHAPTER III

PROCEDURES

Subject Selection

The subject for this study was chosen from the two reported cases of Prader-Willi Syndrome in the geographic area. The subject was enrolled in a 24-hour residential facility and received direct service delivery in adapted physical education by the investigator.

Treatments

Interventions were programmed by trained professionals at the Georgia Retardation Cener - Athens Unit in, but not limited to, the following services: adapted physical education, behavior modification, nutrition, nursing, dormitory living, social work, education, and speech and language. The following is a description of the various intervention strategies.

Adapted physical education: The physical education programs prescribed for the subject were exercised-based utilizing the bicycle ergometer, treadmill, swimming pool, and the gymnasium. Physical fitness was targeted as the primary need. The subject was put on the program at a progressive rate. She started at one mile on the bicycle ergometer and, at last report, was up to five miles without stopping. Swimming was used not only as an exercise activity, but also as a reinforcer for appropriate behavior. Traditional exercises and walking/jogging were prescribed to increase mobility, muscular strength/endurance, and cardiovascular/cardiorespiratory endurance.

Behavior modification: The treatment procedure involved a simple behavioral exchange in which the subject exchanged good behavior for certain activities or privileges. Problem areas that were identified and targeted for change were incorporated into a "YES-NO" type question, where a "YES" response from a staff member indicated that the subject's behavior moved in the desired direction. When the subject received the set number of "YES" responses, she was immediately rewarded with the stated activity or privilege. This type of intervention was identified as a modified form of the Premack Principle. The daily forms with the questions served to remind the subject that her behavior was related to receiving a reward, yet provided documentation of her compliance with staff requests (see Appendix E). The subject was also treated by drug therapy. She was on Ritalin for control of behavior; however, due to lethargy, it was discontinued and substituted with Dexadrine.

Nutrition: The subject's diet when she entered the Georgia Retardation Center was for 1800 Kcal which represented her recommended daily allowance for calories for her height and age. At this point, she continued to gain weight. Although her diet had been controlled at mealtime at the Center, some deviations were reported while she was engaged in activities away from the Center. Based on the continued weight gain, the subject's food intake was reduced to a 1500 Kcal diet. Diet therapy and counseling help the subject find the adjustive resources to cope with the degree of stress associated with her condition and the preoccupation with food.

Nursing: The nursing care plan for the subject consisted of three main objectives. First, the subject received preventive intervention to detect any medical problems. Second, she received preventive dental

intervention that would promote good dental health and hygiene. Last, the subject would maintain good nutrition and the desired height and weight for her age as accurately as possible in consideration of Prader-Willi Syndrome characteristics.

Dormitory living: The dormitory staff continues to provide consistent reinforcement of treatments following the school day.

Recreation: Therapeutic recreation consisted of group and individual play. The subject was taken into the community as much as possible to train her in appropriate socialization skills.

Social work: The planned intervention for the subject's social work plan demonstrated four basic needs: (1) a less restrictive community residential placement; (2) parent training to learn behavior management techniques; (3) regular contact with her family; and, (4) to develop more internal control of her behavior. In conjunction with these needs, four plans were developed and implemented: (1) the subject's parents, school, and developmental service representative were kept aware of her progress, problems, programs, and needs; (2) her parents were requested to participate in parent training to learn how to implement her behavior management plan at home; (3) contact between the subject and her family was encouraged through visits to the Center, visits home, and telephone calls; and, (4) the social worker participated in providing rewards for good behavior as written in her behavior management plan.

Education: Since the subject was perhaps the highest functioning student at the Center, she was placed in the Adolescent III classroom. Programming included self-help skills, pre-vocational training, basic educational skills, and behavior modification. The subject worked with

the Center receptionaist in the morning doing miscellaneous jobs such as cleaning and running simple errands. This was done to improve her socialization skills and improve her self-concept.

Speech and language: Due to the subject's adequate communication skills, speech therapy was not recommended for her. Her hearing was reported normal for both ears via an audiological assessment.

Data Collection

Data were collected on the subject in three areas: behavior, weight, and adjustment to physical activity. Non-quantitative data used are located in the following appendices:

Appendix B	Tests of Resting and Exercise Metabolism
Appendix C	Previous Staffing Summaries
Appendix D	IPP Staffing with Detailed Interdisciplinary Input

Analysis of Data

The analysis of data was completed by utilizing the subject's developmental and medical history; continuous evaluations (monthly and quarterly reviews); previous staffing summaries; motor proficiency tests; an exercise metabolism test; nursing reports; the IPP; interdisciplinary progress reports; the behavior management plan; other treatment team meetings; and, a parent questionnaire.

Consent Forms

A sample of the consent form and approval of the study are contained in Appendix A. The consent form was signed by the parent and is on file with the investigator. Approvals were obtained from the Department of Human Resources and the University of Georgia.

CHAPTER IV

RESULTS AND DISCUSSION

The subject for this study was a 15 year old caucasian female. She was born to unrelated parents 3½ weeks early. She could not suck at birth and was fed by a dropper. She was fed two ounces every two hours for four months. Each feeding lasted approximately 45 minutes.

The mother has been pregnant twice and both resulted in live births. An older brother, aged 16, has no health or physical problems. The mother reported that during the pregnancy, the subject was not very active (poor fetal vigor). After being born 3½ weeks early, the baby had difficulty in breathing and feet turned inward. The baby did not cry immediately, and when stimulated, emitted weak sounds. The subject was not discharged with the mother due to a drop in birth weight from five pounds four ounces to four pounds eleven ounces. It was reported the baby did not cry or suck until one year of age.

The subject maintained a very low weight in her first year. This was due to the absence of the sucking reflex. It was not until three years of age until she could eat table food. She ate great amounts and demonstrated an nsatiable appetite. At 3½ years, the subject was admitted to the Scottish Rite Hospital. Additional tests and observations were made at the Emory Clinic. At this point, the subject was diagnosed as having many characteristics of the classic Prader-Willi individual.

At the age of five, the mother reported behavior problems resulting from the thwarting of food demands. Such episodes as stealing food,

drinking fluids that others had left behind, and others occurred. At age 14, the subject was referred to the Georgia Retardation Center - Athens Unit by her parent and the Clayton Mental Health/Mental Retardation Center for residential treatment for weight loss and behavior modification.

As the etiology of Prader-Willi Syndrome is unknown, reports range from chromosome damage to hypothalamic dysfunction. At one year of age, the subject was tested for chromosome damage, with none found. There is no evidence of mental retardation, blood disease, or any unexplained deaths in the family history.

The child's development was delayed. Crawling was accomplished at 20 months; standing at 26 months; walking at 36 months; words at 24 months; and, sentences at 4 years. Dressing skills were mastered at ages 4 - 5.

Present Level of Functioning

At the subject's IPP staffing (approximately one month after admission at the Center) the subject's present level of functioning, needs, and goals and objectives were stated. The subject's diagnosis was mild mental retardation, exogenous obesity, compulsive eating, small hands and feet, aggressive behavior, unsocialized behavior, and hyperkinesis. She was admitted to the Center for behavior management and weight control.

The subject was evaluated or assessed by all disciplines and the following was reported:

Perceptual Skills: The subject has the perceptual skills to respond effectively with her environment.

Sensori-motor Skills: The subject showed strengths in catching, throwing, and striking; whereas, weaknesses were running, hopping, and balance. No physical limitations

or handicaps were noted.

Self-help Skills: The subject independently performs skills needed for daily personal care.

Communication Skills: Her spontaneous speech is readily intelligible; and, both her receptive and expressive language were adequate for her level of functioning.

Social Skills: The subject has no peer group in her current placement. She communicates with the staff, but the students are non-verbal on her dormitory floor; therefore, the subject shows impatience at times. She demonstrates unacceptable behavior of flopping and closing eyes when being talked to by staff members; although, she responds well to praise.

Emotional Stability: The subject demonstrates some behaviors which indicate a degree of emotional instability. She quickly alters her mood from cooperative to uncooperative with little or no provocation. She also demonstrates many manipulative and counterproductive behaviors that are caused by her environment.

Self-direction: She can initiate tasks and does so if the task is a favorable one. It does not appear she can move from one task to another without redirection.

Effective Use of Time: The subject's mood seems to dictate how she chooses to use her leisure time.

The subject's strengths, weaknesses or problems, and needs were

identified as follows:

Strengths

1. Intelligible speech
2. Self-care skills
3. No physical disability other than Prader-Willi Syndrome
4. Good cognitive ability
5. Vision and hearing appear to be adequate

Problems

1. Non-compliant behavior and temper tantrums
2. Obesity and an over-whelming preoccupation with food
3. Physical activity level
4. Picking at sores

Needs

1. Behavior management plan
2. Weight control
3. Increased physical activity
4. Medical intervention

Two major goals were developed for the subject:

1. To exhibit more compliant, cooperative behaviors as indicated through data recorded for behavior management plan.
2. To decrease weight through a calorie-controlled diet and increase physical activity.

At the last reported date, the subject was making progress on both goals. She is now attending classes in the Clarke County School System and residing at the Georgia Retardation Center - Athens Unit. This

alternative placement for part of the day seems to enhance the subject's self-concept and provide her with the necessary personal interaction with peers.

Comparison of Subject with Current Literature Findings

Since the etiology of Prader-Willi Syndrome appears not to be attributed to any single factor, the investigator suggests multi-factorial causation of possible unknown teratogens is the cause of the syndrome in the subject, which concurs with Bonuccelli et al. (1982). All cases reported have similar characteristics but they are not consistent throughout the Prader-Willi population.

According to many studies, a possible etiology of Prader-Willi Syndrome is chromosomal abnormalities (abberations/translocations/deletions). However, at one year of age, the subject was tested for chromosomal abnormalities and none were found. Therefore, the subject does not fall into the approximated ten percent of Prader-Willi individuals who have some type of chromosomal abnormality.

The characteristics that the subject demonstrates that are considered classic to the Prader-Willi individual are: poor fetal vigor (Bray et al., 1983), non-term delivery (Bray et al., 1983), low birth weight (Hoefnagel et al., 1967), hypotonia (Butler et al., 1982), feeding problems (Bonuccelli et al., 1982), delayed milestones (Prader-Willi Syndrome Association, 1981), obesity (all of the literature), short stature (Nardella et al., 1983), insatiable appetite (Otto et al., 1982), lack of balance (Prader-Willi Syndrome Association, 1981), small hands and feet (Prader-Willi Syndrome Association, 1981), and scratching and

picking at sores (Prader-Willi Syndrome Association, 1981). Wannarachue et al. (1975) reported that of the female patients studied, the sexual development varies. The subject of this study has a history of menstruation. The subject has normal external genitalia and the menstruation has consisted of two scanty periods.

In concurrence with Otto et al. (1982), Prader/Willi Syndrome Parents & Friends (1977), and Thompson et al. (1980), the subject demonstrates polyphagia, characterized by continuous eating and an insatiable appetite. The subject has also demonstrated temper tantrums, moodiness, stealing food, and erratic behavior.

The subject was tested in the Human Performance Laboratory at the University of Georgia on resting and exercise metabolic rates. Her resting metabolic rate was typical for an adult under the testing conditions. However, the results could not be compared with basal metabolic rate standards since she was not in the basal condition during the testing. It was concluded that the subject is likely to have a "very low physical work capacity (maximal oxygen uptake)", although it could not be measured or estimated from the data collected (see Appendix B for the complete report). As the subject is purported to possess a low physical work capacity concurring with Carman's study (1981), then a perpetuating cycle of inactivity leads to low energy output; which in turn leads to a reduction in muscle strength coupled with the continuous desire for food. All other factors tested that were found to be normal for an adult under the testing conditions dispute Bray's et al. (1983) findings of marked reduction in cardiorespiratory factors.

Progress of Subject

According to data collected, the subject has made considerable progress since her admission to the Georgia Retardation Center on April 14, 1983. Non-compliant behavior decreased from a 50 percent weekly average to a 2 percent weekly average in three months (see figure 1). In compliance versus non-compliance for a specified seven day period, the subject demonstrated seven non-compliant behaviors the first day; then, the non-compliant behaviors decreased to an average of one on the second day and remained constant for the remainder of the observation period (see figure 2). The subject demonstrated six compliant behaviors the first day; then, the compliant behaviors increased to an average of thirteen per day for the remainder of the observation period (see figure 3). Non-compliant behaviors were observed for a nine day period approximately one month after the subject's admission to the Center. There were a total of 51 acts of non-compliance for an average of 5.67 acts per classroom day (see figure 4). Flopping to the floor was identified as one of the targeted behaviors of the subject. Incidents of flopping for four weeks were documented upon the subject's admission to the Center. The subject demonstrated a total of five flops in the classroom and six flops on the dorm (see figure 5).

The subject's weight has decreased from 175½ pounds upon admission to 161½ pounds at the last reported weighing (see figure 6 for a month by month chart). The subject was tested using skinfold calipers at the Human Performance Laboratory. The subject's tricep skinfold measurement was 30 mm. That places her below the 5th percentile (indicating overfatness) for girls her age in the United States.

FIGURE 1

WEEKLY AVERAGE OF NON-COMPLIANCE

OVER A 3 MONTH PERIOD

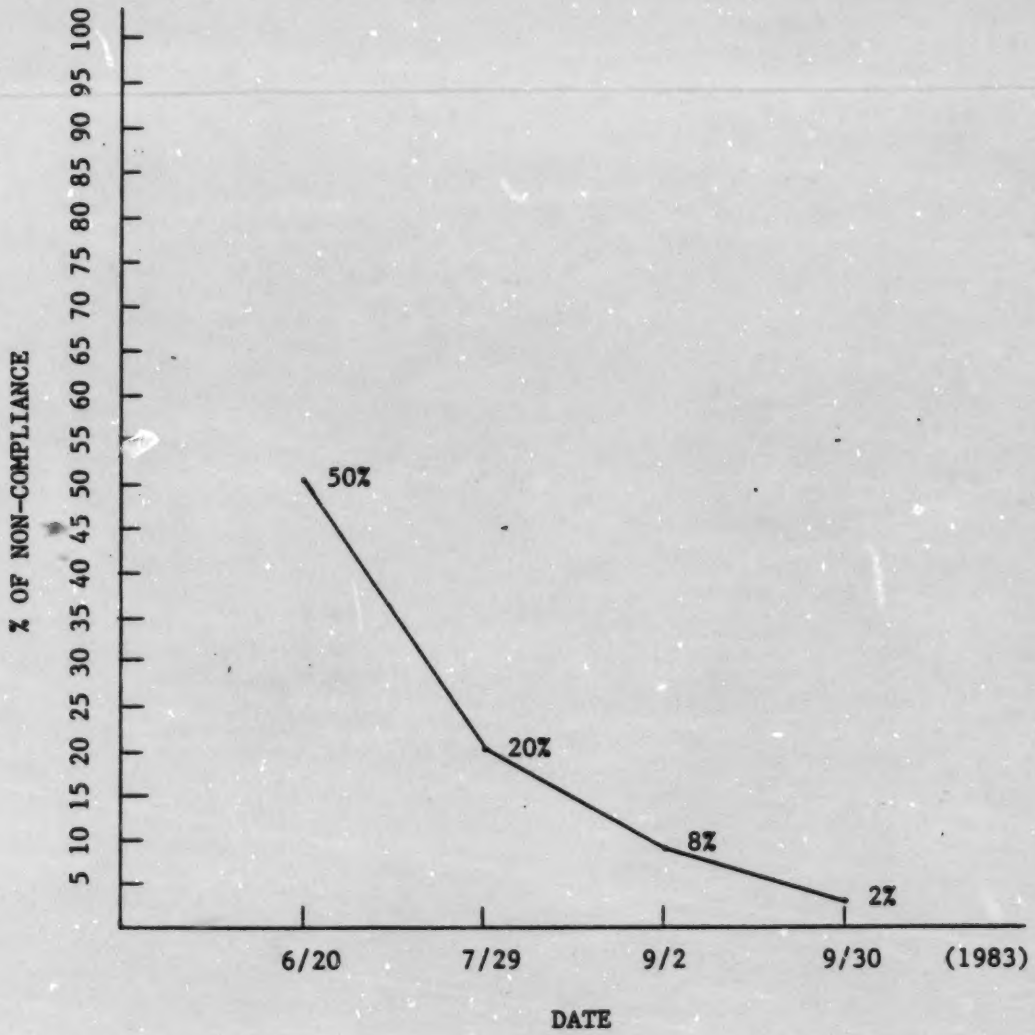
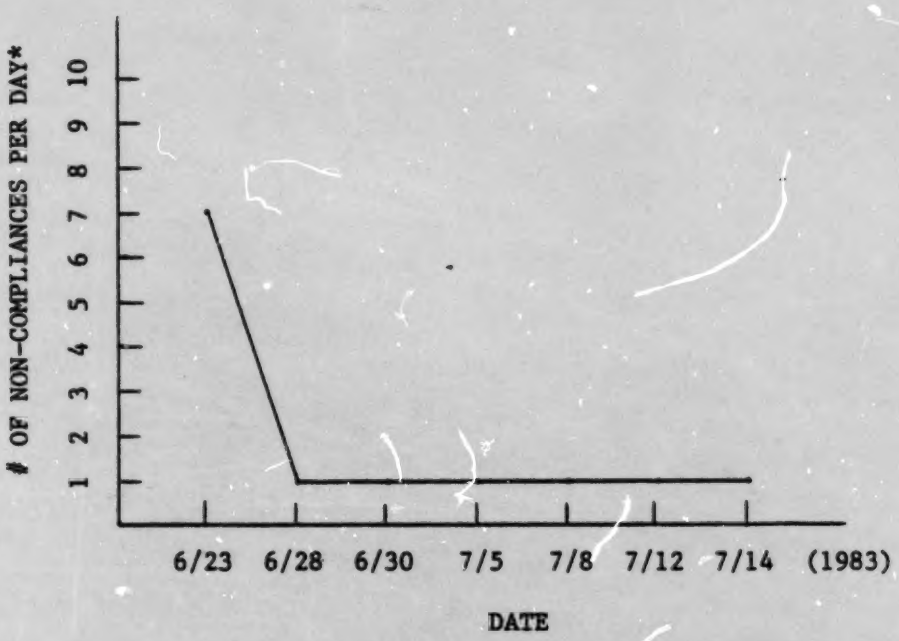


FIGURE 2

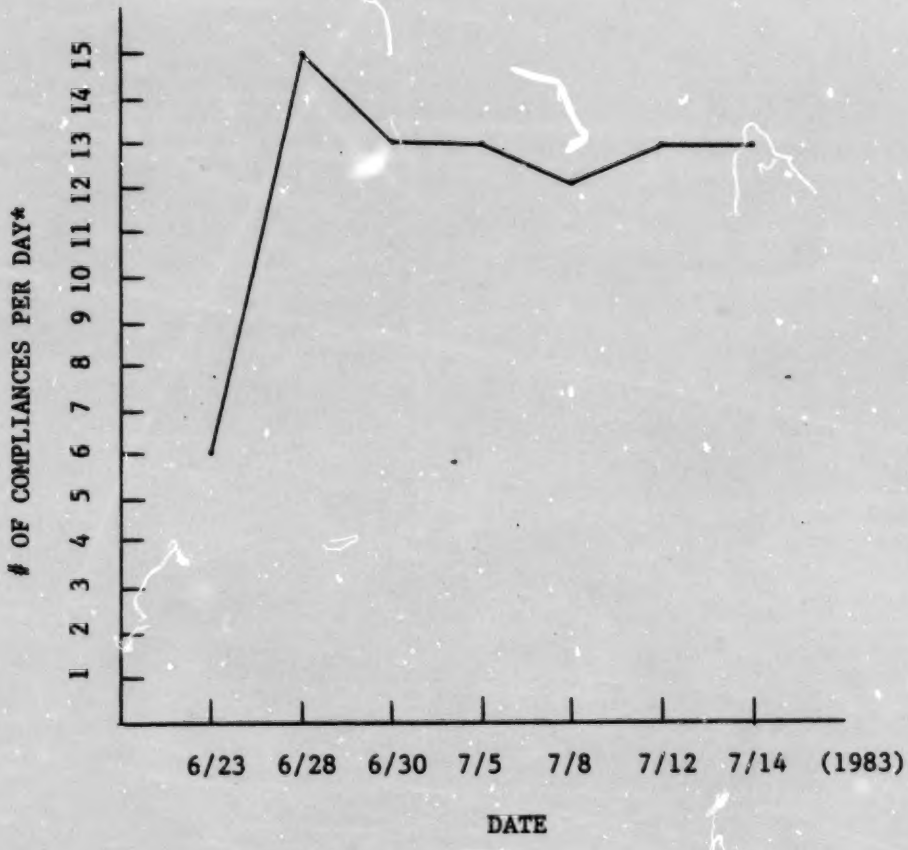
NUMBER OF NON-COMPLIANCES PER DAY



* "per day" = the hour the subject was observed

FIGURE 3

NUMBER OF COMPLIANCES PER DAY



* "per day" = the hour the subject was observed

FIGURE 4

NON-COMPLIANCE PER CLASSROOM DAY

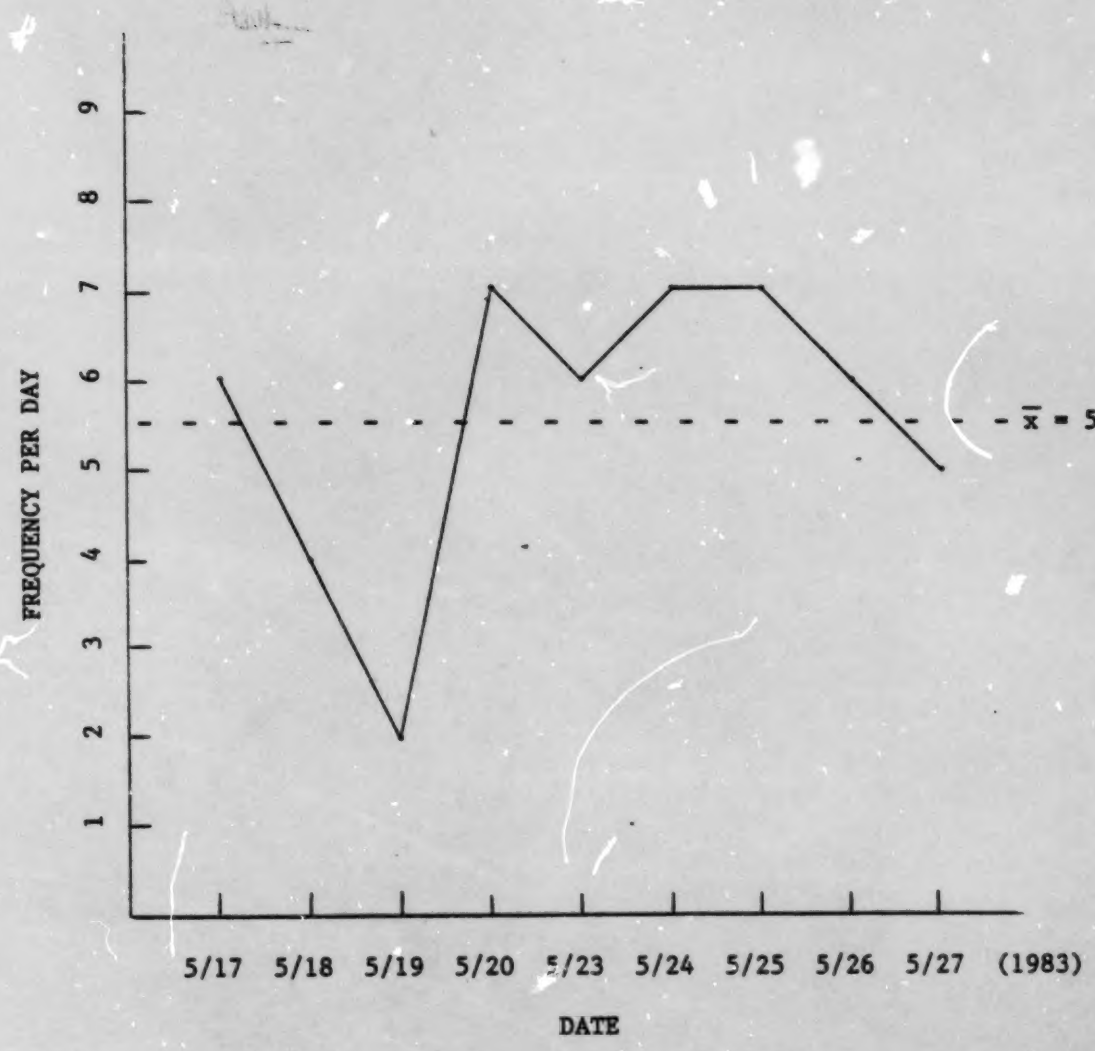


FIGURE 5

FREQUENCY OF FLOOR FLOPPING

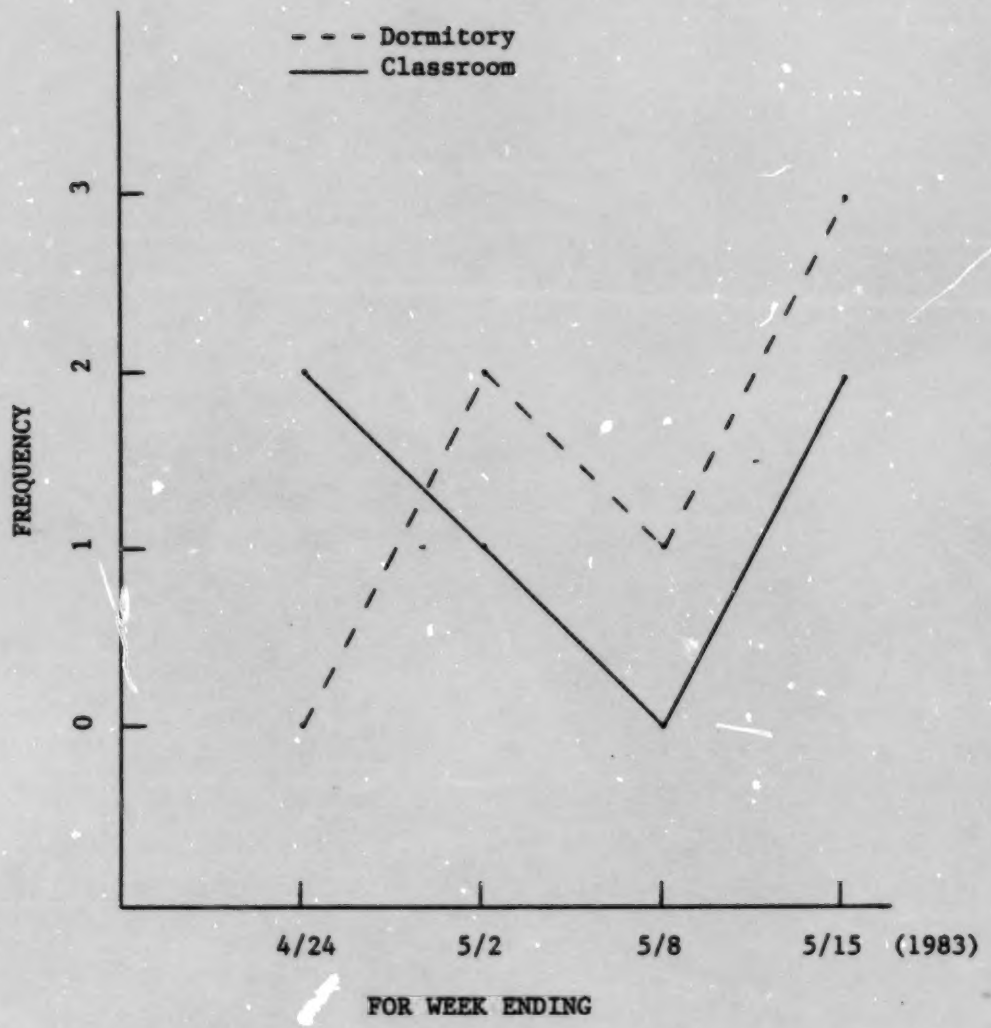
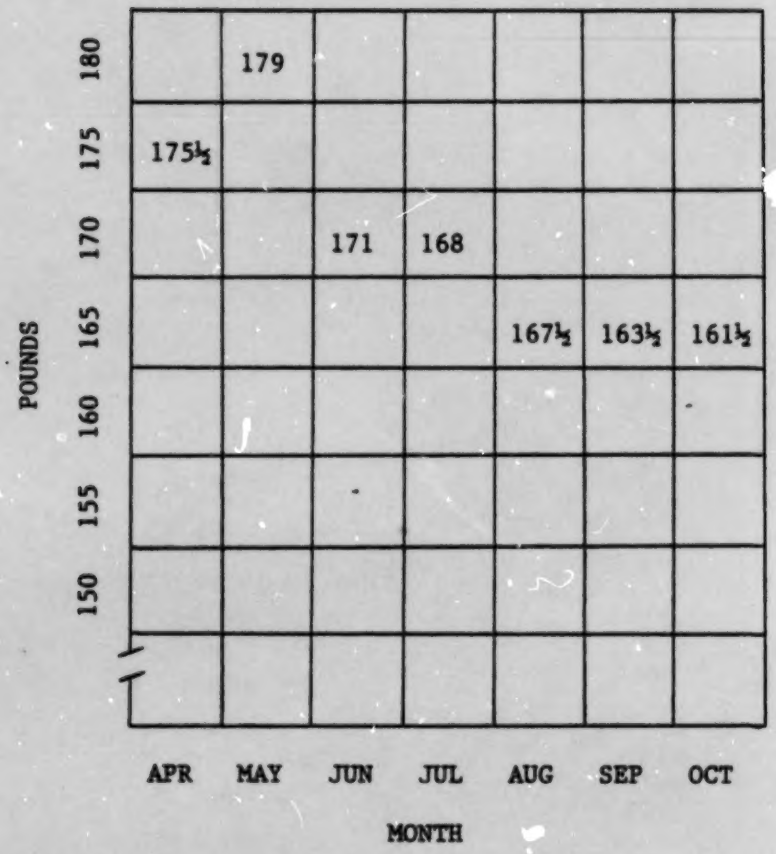


FIGURE 6

WEIGHT CHART



The Seefeldt Selected Gross Motor Skills test (a modified version) was used to assess the subject on motor proficiency upon admission and administered again six months later. A copy of this instrument appears in Appendix F. The subject improved on two of the six itmes that were tested and remained unchanged on the other four (see figures 7 & 8).

Placing the subject in the Clarke County School System has proven to be very beneficial to the subject. However, no data are presently available on her progress. Reportedly, she is able to interact with peers and interact within the community.

FIGURE 7

EVALUATION FORM FOR SELECTED GROSS MOTOR SKILLS

SKILL	1	2	3	4	5	COMMENTS:
Running and Walking			X			
Hopping	X					Low due to balance
Kicking				X		
Striking				X		
Throwing				X		Relative strength; needs some work
Catching			X			Relative strength; needs some work

William R. Forbus, A. P. E.

April 25, 1983
Date

FIGURE 8

EVALUATION FORM FOR SELECTED GROSS MOTOR SKILLS

SKILL	1	2	3	4	5	COMMENTS:
Running and Walking				X		
Hopping			X			Low due to balance
Kicking				X		
Striking				X		
Throwing				X		Relative strength; needs some work
Catching			X			Relative strength; needs some work

William R. Forbus, A. P. E.

October 14, 1983
Date

CHAPTER V

SUMMARY, CONCLUSIONS AND RECOMMENDATIONS

Summary

The purpose of this study was to investigate and chronolog the status and experiences of a diagnosed Prader-Willi child, and to determine how the child compares with current research and literature. This was accomplished by reviewing the current research and literature and analyzing data from the subject's developmental and medical histories, continuous evaluations, previous staffing summaries, motor proficiency tests, an exercise metabolism test, nursing reports, the IPP, inter-disciplinary progress reports, the behavior management plan, treatment team meetings, and a parent questionnaire.

The results of the study revealed that the subject demonstrates most of the characteristics that are classic to the Prader-Willi population. The subject has also made considerable progress toward reaching her program goals.

Conclusions

In consideration of the purpose and the outcome of the study, the following conclusions appear warranted:

1. The subject does demonstrate most of the characteristics that are classic to the Prader-Willi population and her programming should encompass her problem areas.

2. The subject should be placed close to the mainstream due to her present level of functioning, capabilities, and capacities.

3. The subject has shown improvement in all three interventions (physical activity, nutrition counseling, & behavior modification) and, therefore, these should continue.

Recommendations

The results of this study have suggested recommendations specific to the programming of the child:

1. Continue intervention strategies in physical activity, nutrition counseling, and behavior modification.

2. Assess the subject using the Bruininks-Oseretsky Test of Motor Proficiency and continue monitoring the physiological parameters of the child.

3. Test the subject for chromosome abnormalities.

The development of this study has suggested possibilities for further investigations. Among the recommendations are:

1. A study comparing the subject with another diagnosed Prader-Willi child.

2. A study concerned with the child's basal metabolic rate and the possibility of the child expending fewer calories than the normal child.

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APPENDICES

APPENDIX A

INFORMED CONSENT FORM

RESEARCH APPROVAL

INFORMED CONSENT FORM

I, the undersigned, give consent for _____ to participate in the research project entitled PRADER-WILLI DISEASE: A CASE STUDY, which is being conducted by WILLIAM R. FORBUS, III. I understand that this participation is entirely voluntary; I can withdraw my consent at any time and have the results of the participation returned to me, removed from the research records, or destroyed.

The following points have been explained to me (or my child):

1) The reason for the research is to investigate the parameters encompassing a Prader-Willi child's life style and extrapolate the effects of a multi-disciplinary approach on the child's present level of functioning and future physical/social status, and the benefits that I may expect from it are information and possible treatment techniques to use with Prader-Willi children.

2) The procedures are as follows: The subject's medical history, present level of functioning and interdisciplinary interventions (especially physical education) will be examined by family interviews, review of medical records, review of current literature and research, interview with a cytogeneticist, and study of present functioning level in physical education using a motor proficiency test.

3) No discomforts or stress will be faced during this project.

4) Participation entails no risks.

5) The results of this participation will be confidential and will not be released in any individually identifiable form without my prior consent unless required by law.

6) The investigator will answer any further questions about the research, either now or during the course of this project.

_____ _____

William R. Forbus date Legally Authorized Representative
Investigator

PLEASE SIGN BOTH COPIES. KEEP ONE AND RETURN THE OTHER TO THE INVESTIGATOR.

Research programs at the University of Georgia which involve human subjects are carried out under the oversight of the Institutional Review Board. Questions or problems regarding these activities should be addressed to Dr. Nathan W. Dean, Assistant Vice-President for Research, 608 Graduate Studies Research Center, (542-3360).

THE UNIVERSITY OF GEORGIA
1785 • BICENTENNIAL • 1985

OFFICE OF THE VICE PRESIDENT FOR RESEARCH

July 29, 1983

Mr. William R. Forbus, III
850 College Station Road
Campus

Dear Mr. Forbus:

Thank you for your study, "Prader-Willi Disease: A Case Study." It has been approved through November 1, 1983, as per your request.

Please let me know if there are any additions or changes in your study or if I can be of any further help.

Sincerely,

Pamela Wier

Pamela Wier
Executive Secretary
Institutional Review Board

cc: Dr. Ernest Bundschuh

enclosure

BOYD GRADUATE STUDIES RESEARCH CENTER
THE UNIVERSITY OF GEORGIA
ATHENS, GEORGIA 30602
(404) 542-3360

AN EQUAL OPPORTUNITY / AFFIRMATIVE ACTION INSTITUTION

James G. Ledbetter, Ph.D./Commissioner

GEORGIA DEPARTMENT OF HUMAN RESOURCES

878 PEACHTREE STREET, N.E. / ATLANTA, GEORGIA 30309

August 15, 1983

M E M O R A N D U M

TO: W. R. Forbus, III
777 Sunnyside Drive
Athens, Georgia 30606

FROM: Greg Peterson, Executive Secretary
DHR Human Research Review Board

SUBJECT: Research Proposal: "Prader-Willi Disease"

The subject proposal has been reviewed and approved by the Review Board. You are authorized to undertake the research in accordance with the provisions of the proposal.

cc: Mr. Bill Riddle
Unit Director
GRC-Athens
850 College Station Road
Athens, Georgia 30610

APPENDIX B

TESTS OF RESTING AND EXERCISE METABOLISM

THE UNIVERSITY OF GEORGIA

Inter-office Communication

To: Dr. Ernie Bundschuh Date: September 7, 1983

From: Kirk Cureton KJC Phone: 2-8782

Subject: Tests of Resting and Exercise Metabolism

This is a report on the data collected on on September 2, 1983. Her metabolic rate was measured using open circuit spirometry at rest in the supine position following a 15-minute period of lying relatively still and, during bicycle ergometer exercise performed for 3 minutes at 0 kp and 50 rev/min. Attempts to have her exercise at a higher work rate were unsuccessful. An ECG was also taken at rest and during exercise.

Data collected is summarized below.

Height	144.8 cm	56.7 in
Weight	73.9 kg	162.6 lb
Triceps skinfold	30 mm	

	Rest	Exercise
Ventilation (l/min)	8.0	16.6
Oxygen uptake (l/min)	0.233	0.497
Oxygen uptake (ml/kg/min)	3.1	6.7
Energy expenditure (Kcals/min)	1.16	2.41
Carbon dioxide production (l/min)	0.225	0.423
Respiratory exchange ratio	0.97	0.85
Heart rate (b/min)	77	100

The resting metabolic rate obtained is typical for an adult under these conditions. The value cannot be compared with basal metabolic standards since she was not in the basal condition. Her respiratory exchange ratio indicates she hyperventilated somewhat during the time which is not unusual for someone slightly anxious. The resting heart rate is normal.

During the exercise, her metabolic rate was approximately doubled. This was still a very low level of exercise stress as indicated by her heart rate. She apparently is unwilling to tolerate even slight discomfort since when the work rate was increased to 0.25 kp at 50 rev/min, a very small increase, she immediately expressed displeasure and refused to maintain the pedal rate. It is likely she has a very low physical work capacity (maximal oxygen uptake), but we could not measure or estimate it from the data collected. It is

possible a treadmill test would yield more useful data.

There was no indication of an abnormality in her ECG that Dr. DuVal detected. I have enclosed the resting ECG in case you wish to keep it in her file.

Her triceps skinfold was below the 5th percentile (indicating overfatness) for girls her age in the U.S.

I hope this information will be of some value.

KC/eh
Enclosure

APPENDIX C

PREVIOUS STAFFING SUMMARIES

ALL PREVIOUS STAFFING SUMMARIES CAN BE REVIEWED
IN THE MEDICAL RECORDS SECTION AT THE:

Athens Unit GRC
850 College Station Road
Athens, GA 30610

APPENDIX D

IPP STAFFING

WITH

DETAILED INTERDISCIPLINARY INPUT

BEST COPY

GEORGIA RETARDATION CENTER - ATHENS

IPP/STAFFING

NAME: Parent/Legal Guardian:
BD: 03-09-68
SS:
Date of Admission: 04-14-83
Date of Meeting: 05-12-83

did not attend her IPP because the treatment team felt she would not benefit from participating in this meeting.

Diagnosis:
1. Mental Retardation, Mild, associated with Prader-Willis Syndrome.
   A. Exogenous obesity
   B. Compuslive eating
   C. Small hands and feet
2. Aggressive and unsocialized behavior
3. Hyperkinesis
4. Monitial Vaginitis

Date and Reason for Admission:
, a fifteen year old white female from Clayton County, was admitted to the GRC-Athens Unit on April 14, 1983 for behavior management and weight control, which her mother is unable to currently provide at home.

Assessment Results:
Perceptual Skills - has the perceptual skills to respond effectively with her environment.

Sensori-motor Skills - shows strengths in catching, throwing, and striking. Weaknesses are noted in running, hopping, and balance. does not like to perform most assembly type work tasks, but there does not appear to be any physical reason to prevent her from performing work activity tasks.

Self-Help - independently performs the self-help skills needed for daily personal care.

Communication - spontaneous speech is readily intelligible. Both her expressive and receptive language are adequate for her level of functioning.

Social Skills - does not have a peer group in her current placement. She communicates with staff, but the students on her dorm are non verbal, and seems to lose patience with them at times. displays some immature behaviors with staff; such as flopping to the floor and closing her eyes when talking with adults. usually responds well to praise from others. Special attention from adults can be used as a reward for good behavior.

BEST COPY

BD: 03-09-68
SS:

Emotional Stability - demonstrates some behaviors which indicate a degree of emotional instability. She quickly alters her mood from cooperative to non-cooperative with little or no provocation approximately 3 times a day. She also demonstrates many manipulative and counterproductive behaviors which are caused by her environment or situation.

Self-direction - can initiate tasks and does so if she wants to perform the task. It does not appear that can move from a task once it is completed to another task.

Effective Use of Time - mood seems to determine how she chooses to use her time. She is working on a latch hook project, but also spends much of her free time sleeping.

Statement of Strengths, Problems and Needs:

A. Strengths
1. Intelligible speech and communicates in complete sentences.
2. Self-care skills
3. No physical disabilities other than Prader-Willi Syndrome
4. Good cognitive ability for functioning level
5. Vision and hearing appear to be adequate

B. Problems
1. Non-compliant behaviors and temper tantrums
2. Obesity and an overwhelming pre-occupation with food
3. Physical activity level
4. Picking at sores

C. Needs
1. Behavior management for non-compliant behaviors and temper tantrums
2. Weight control - low calorie diet
3. Increased physical activity - adapted physical education
4. Frequent monitoring of skin irritation areas to determine need for medical intervention.

Statement of Goals (See attached Goal/Objective Sheet for Objectives):

A. will exhibit more compliant, co-operative behaviors as indicated through data recorded for behavior management plan.

B. weight will decrease weekly over the next 3 months from her current weight of 177 pounds, through a calorie controlled diet and increased physical exercise.

Prognosis:

It is the opinion of the treatment team that the above stated goals and objectives are appropriate to address reason for admission and that she will accomplish these goals.

program will be monitored on a monthly basis by her QMRP and adjustments made as needed. The full treatment team will review progress on a quarterly basis.

- CONTINUED -

BEST COPY

BD: 03-09-68
SS: - -

Service Recommendations:
1. Request placement for in local YWCO Girl's Summer Program.
2. If remains at GRC-Athens Unit past August 1983, request public school placement.
3. weight and effectiveness of calorie controlled diet to be monitored weekly.
4. Increased exercise - adapted physical education
5. Reassessment of medication at next quarterly review
6. Parent training in behavior management techniques

Behavior Management Plan:
A preliminary behavior management plan has been designed and should be implemented within a week. The plan includes a combination of counseling and behavior management techniques.

Criteria for Release/Least Restrictive Environment/Alternative Placement/ Projected Length of Stay:
treatment team agrees that placement for at GRC-Athens Unit with the current population of Severely and Profoundly retarded students is not the least restrictive environment in which her needs can be met. However, at this time, mother cannot cope with behaviors and an alternative community placement is not available. Considering the only current option of placing back in the home, GRC-Athens Unit is presently the least restrictive available environment in which needs for behavior management and weight control can be met.

will be released from GRC when her non-compliant behaviors and temper tantrums have been reduced 50% from baseline data through an effective behavior management plan which can be implemented by her mother, school teachers, and babysitters; and when she has shown a steady decline in her weight over a three month period.

Though projected length of stay is until 04-14-84, her program will be monitored quarterly and she will be released as soon as she meets her criteria for release or a less restrictive community placement can be found for her.

Statement of Human Rights, Custody and Need for Guardianship:
is a minor child who is unable to fully understand, exercise, and protect her human rights. Her natural mother, , is her guardian. When was admitted to GRC-Athens Unit, her rights were explained and discussed with her mother. While a student at GRC - rights will be protected in all areas of programming.

Additional Reports and Evaluations available in students permanent record:
Social Work, Psychological, Speech, Physical Education, Nutrition, Nursing/ Medical, Behavior Management

- CONTINUED -

Name _____
BD _____
SS# _____

Date _____

Goal	Objective	Initiated Date	Target Date	Responsible Person	Method of Evaluation	Rev Dat
Mealtime Program	self-feeds; however she needs reminders to slow down and put the fork down after each bite.	6-83		Conrado, Classroom Autry, dorm		grad

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DD 83-09-09

SS#

Goal	New Objectives	Initiated Date	Target Date	Responsible Person	Method of Evaluation	Rev Dat
A. will exhibit more compliant, co-operative behavior as indicated through data recorded for behavior management plan. (Same as on old Goal/ Objective sheet)	A1. will receive the maximum amount of "Yes"es for a period of 1 day. (Refer to behavior management plan for detail)	09-10-83	09-31-83	Linda Hagan QMRP	Review complied Behavior Management data collection forms	Mon
	A2. will receive the maximum amount of "Yes"es for a period of 3 days.	When A1 is Achieved	N/A	"	"	
	A3. will receive the maximum amount of "Yes"es for a period of one week.	When A2 is achieved	N/A	"	"	"
B. weight will decrease weekly over the next 3 months from her current weight of 177 pounds, through a calorie controlled diet and increased physical exercise.	B1. weight will decrease weekly for a 3 month period.	05-09-83	08-31-83	Thompson, Nursing	Data - 1 weekly weight-ins at approx-mately same time of day with similar clothing.	wee

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SS#

Goal	Objective	Initiated Date	Target Date	Responsible Person	Method of Evaluation	R D
A. will exhibit more compliant, co-operative behavior as indicated through data recorded for behavior management plan.	A1. will receive the maximum amount of signature for a period of 1 day. (Refer to behavior management plan for detail.)	05-30-83	06-27-83	Tiller, Sr. Behavior Specialist	data collection	Mon
	A2. will receive the maximum amount of signatures for a period of 3 days.	When A.1. is achieved	07-25-83	"	"	"
	A3. will receive the maximum amount of signatures for a period of one week.	When A.2. is achieved	08-31-83	"	"	"
B. weight will decrease weekly over the next 3 months from her current weight of 177 pounds, through a calorie controlled diet and increased physical exercise.	B1. weight will decrease weekly for a 3 month period.	05-09-83	08-31-83	Thompson, Nursing	Data -1 weekly weight-ins at approximately same time of day with similiar clothing.	we
	B2. will put her fork down between every bite.	05-23-83	08-31-83	Dorm Staff	Observation/ Data	Mon
	B3. will walk 2 laps around the parking lot at least 3 times a week.	05-23-83	08-31-83	PE Staff	"	"

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APPENDIX E

BEHAVIOR MODIFICATION CHECKLIST

DAILY CHECKLIST

DATE:_____

Directions: Circle either YES or NO as indicated by the subject's behavior, and initial at the side. This form will follow the subject throughout the day and will be stored on fourth floor at the end of the day.

WAKE UP TIME UNTIL 8:30 A.M.

1. Comes to breakfast on time, leaves the floor with the other students and arrives at the lunchroom with the other students not more than 15 seconds after the last student is through the lunchroom door.

YES NO

2. Finishes breakfast when the other students do and is ready to leave the lunchroom when they are.

YES NO

3. Returns to the dorm in a mature fashion with no intervention or verbal direction needed from staff. No more than one verbal command is needed and she arrives in the dorm area no more than 15 seconds after the last person.

YES NO

4. Comes to class on time and no later than 8:30 A.M. unless specifically excused by staff.

YES NO

Total YESes = (Maximum this section = 4)

If YESes = 4, then the subject can collect attendance slips and work with the receptionist or receive some other privilege as designated by the teacher.

AFTER SCHOOL TO DINNER

1. Stays in room or activity room during quiet time.

YES NO

2. Gets up from nap when called (if applicable).

YES NO

Total YESes = (maximum this section = 2)

If YESes = 2, then the subject can watch a TV program of her choice.

DINNER TIME UNTIL BEDTIME

1. Returns to dorm after dinner under same conditions as the breakfast rule (section 1, number 3).

YES NO

2. Takes bath and washes hair in the time designated by staff and not exceeding 30 minutes.

YES NO

Total YESes = (Maximum this section = 2)

If YESes = 2, then the subject can stay up until 10:00 P.M. in order to work on hobbies or watch TV.

GENERAL DAILY RULES

Directions: Circle YES or NO as applicable in your program area and initial.

CLASSROOM:
1. Does not fall to floor at any time during the classroom day.

YES NO

DORMITORY:
2. Did not fall to floor at any time during dorm hours today.

YES NO

3. Followed special instructions of the dormitory staff in regard to helping in the dorm.

YES NO

4. Has not bothered any other students during any part of the day. This means that she <u>does not</u> go into their rooms in the morning, remove their clothing, comb their hair or redress them or try to work with them in any way.

YES NO

5. Goes where directed by staff and remains there until told to move somewhere else.

YES NO

Total YESes = (max. per day Mon.-Fri. only = 5)

If the subject has 25 YESes at the end of the week, she will receive a special treat arranged by the staff.

APPENDIX F

SEEFELDT'S SELECTED GROSS MOTOR SKILLS TEST

Developmental Sequence of Catching

Stage 1. The child presents his arms directly in front of him, with elbows extended and the palms facing upward or inward toward the mid saggital plane. As the ball contacts the hands or arms the elbows are flexed and the arms and hands attempt to secure the ball by holding it against the chest.

Stage 2. The child prepares to receive the object with the arms in front of the body, the elbows extended or slightly flexed. Upon presentation of the ball the arms begin an encircling motion which culminates by securing the ball against the chest. Stage two also differs from stage one in that the receiver initiates the arm action prior to ball-arm contact in stage two.

Stage 3. The child prepares to receive the ball with arms which are slightly flexed and extended forward at the shoulder. Many children also receive the ball with arms which are flexed at the elbow, with the elbow ahead of a frontal plane.

Substage 1. The child uses his chest as the first contact point of the ball and attempts to secure the ball by holding it to his chest with the hands and arms.

Substage 2. The child attempts to catch the ball with his hands. Upon his failure to hold it securely he maneuvers it to his chest, where it is controlled by hands and arms.

Stage 4. The child prepares to receive the ball by flexing the elbows and presenting the arms ahead of the frontal plane. Skillful performers may keep the elbows at the sides and flex the arms simultaneously as they bring them forward to meet the ball. The ball is caught with the hands, without making contact with any other body parts.

Stage 5. The same upper segmental action is identical to stage four. In addition, the child is required to change his stationary base in order to receive the ball. Stage five is included because of the apparent difficulty which many children encounter when they are required to move in relation to an approaching object.

Developmental Sequence of Throwing

Stage 1. The throwing motion is essentially posterior-anterior in direction. The feet usually remain stationary during the throw. Infrequently, the performer may step or walk just prior to moving the ball into position for throwing. There is little or no trunk rotation in the most rudimentary pattern at this stage, but those at the point of transition between stages one and two may evoke slight trunk rotation in preparation for the throw and extensive hip and trunk rotation in the "follow-through" phase. In the typical stage one the force for projecting the ball comes from hip flexion, shoulder protraction and elbow extension.

Stage 2. The distinctive feature of this stage is the rotation of the body about an imaginary vertical axis, with the hips, spine and shoulders rotating as one unit. The performer may step forward with either an ipsilateral or contralateral pattern, but the arm is brought forward in a transverse plane. The motion may resemble a "sling" rather than a throw due to the extended arm position during the course of the throw.

Stage 3. The distinctive pattern in stage three is the ipsilateral arm-leg action. The ball is placed into a throwing position above the shoulder by a vertical and posterior motion of the arm at the time that the ipsilateral leg is moving forward. This stage involves little or no rotation of the spine and hips in preparation for the throw. The follow-through phase includes flexion at the hip joint and some trunk rotation toward the side opposite the throwing.

Stage 4. The movement is contralateral, with the leg opposite the throwing arm striding forward as the throwing arm is moved in a vertical and posterior direction during the "wind-up" phase. There is little or no rotation of the hips and spine during the wind-up phase; thus, the motion of the trunk and arm closely resemble those of stages one and three. The stride forward with the contralateral leg provides for a wide base of support and greater stability during the force production phase of the throw.

Stage 5. The "wind-up" phase begins with the throwing hand moving in a downward arc and then backward as the opposite leg moves forward. This concurrent action rotates the hip and spine into position for forceful derotation. As the contralateral foot strikes the surface the hips, spine and shoulder begin derotating in sequence. The contralateral leg begins to extend at the knee, providing an equal and opposite reaction to the throwing arm. The arm opposite the throwing limb also moves forcefully toward the body to assist in the "equal and opposite" reaction.

Developmental Sequence of Striking

Striking with a bat

Stage 1 The motion is primarily posterior-anterior in direction. The movement begins with hip extension and slight spinal extension and retraction of the shoulder on the striking side of the body. The elbows flex fully. The feet remain stationary throughout the movement with the primary force coming from extension of the flexed joints.

Stage 2 The feet may remain stationary or either the right or left foot may receive the weight as the body moves toward the approaching ball. The primary pattern is the unitary rotation of the hip-spinal linkage about an imaginary vertical axis. The forward movement of the bat is in a transverse plane.

Stage 3 The shift of weight to the front-supporting foot occurs in an ipsilateral pattern. The trunk rotation-derotation is decreased markedly in comparison to Stage 2 and the movement of the bat is in an oblique-vertical plane instead of the transverse path as seen in Stage 2.

Stage 4 The transfer of weight in rotation-derotation is in a contralateral pattern. The shift of weight to the forward foot occurs while the bat is still moving backward as the hips and spine and shoulder girdle assume their force-producing positions. At the initiation of the forward movement the bat is kept near the body and elbow extension and the supination-pronation of the hands does not occur until the arms and hands are well forward and ready to extend the lever in preparation to meet the ball. At contact the weight is on the forward foot.

Developmental Sequence of Walking and Running

Stage 1. The arms are extended sideward at shoulder height (high-guard position). The stride is short, and of the shoulder width. The surface contact is made with the entire foot, simultaneously. Little knee flexion is seen. The feet remain near the surface at all time.

Stage 2. Arms are carried at "middle guard" (waist height), the stride is longer and approaches the mid-saggital line. Contact is usually with the entire foot striking the surface simultaneously. Greater knee flexion is noted in the movement of the legs becomes anterior-posterior.

Stage 3. The arms are no longer used primarily for balance. Arms are carried below waist level and may flex and assume a counter-rotary action. The foot contact is "heel-toe". Stride length increases and both feet move along a mid-saggital line. The swing leg flexion may be as great as 90 degrees.

Stage 4. Foot contact is heel-toe at slow or modest velocities but may be entirely on the metatarsal arch during sprint running. Arm action is in direct opposition to leg action. Knee flexion is used to maintain the momentum during the support phase. The swing leg may flex until it is nearly in contact with the buttocks during its recovery phase.

Insufficient movements common to running patterns are: inversion or eversion of the foot during the support phase. Inversion results in a medial rotation of the leg and thigh during the support phase and is characterized by an oblique rather than an anterior-posterior pattern as the leg is brought forward in the swing phase.

Eversion of the foot during the support phase results in lateral rotation of the leg and thigh. This pattern is often accompanied by an exaggerated counter-rotary action of the arms in an attempt to maintain a uniform direction.

Developmental Sequence of Kicking

Stage 1 — The performer is usually stationary with the leg of the striking foot slightly flexed at the knee prior to the foot striking the ball. If the performer moves forward prior to the kick, the steps are short and result as a part of the approach rather than a primary preparation for striking the ball. The thigh of the striking leg is perpendicular to the surface or ahead of the mid-frontal plane as the knee of the striking leg is flexed. The slight knee flexion results in a kick with a pushing action rather than a forceful striking motion. There is little follow-through of the kicking foot, and frequently it is withdrawn from the ball after contact.

Stage 2 — The body is stationary during the initiation of the kicking action. The preparatory motion involves hyperextension at the hip joint and hyperflexion at the knee joint, with the thigh of the striking leg in a position behind the mid-frontal plane. Opposition of upper and lower extremities is present during the kicking motion. The body pivots on the supporting leg, but the force of the kick usually is not sufficient to move the body forward after striking the ball.

Stage 3 — The preparatory phase involves an approach to the ball that includes a deliberate step or series of steps. The striking foot remains near the surface in its approach, indicating a reduction in knee flexion in contrast to stage two. The force of the striking foot is less than maximum, reflected by an upright posture of the trunk. The follow-through may result in the performer moving past the point of contact if the approach was rapid or the performer may remain near the point of contact if the approach was deliberate and cautious.

Stage 4 — The approach to the ball involves one or more steps, but the distance just prior to the kick is covered by a leap. In other words, the kicker must be airborn in the approach to qualify as a stage four kicker. The knee of the kicking leg is slightly flexed by the action of the long leap just prior to kicking. The trunk is inclined backward prior to and during contact, in order to place the rectus femorus muscle in the most efficient position for contraction. The momentum of the kick is dissipated by hopping on the support leg and then stepping in the direction of the object which has been struck.

Developmental Stages of Hopping

Stage 1 The non-support knee is flexed at 90 degrees or less with the non-support thigh parallel to the surface. This position places the non-support foot in front of the body so that it may be used for support in the event that balance is lost. The body is held in an upright position with the arms flexed at the elbows. The hands are held near shoulder height and slightly to the side in a stabilizing position. Force production is generally limited so that little height or distance is achieved in a single hop.

Stage 2 The non-support knee is fully flexed so that the foot is near the buttocks. The thigh of the non-support leg is nearly parallel to the surface. The trunk is flexed at the hip resulting in a slight forward lean. The performer gains considerable height by flexing and extending the joints of the supporting leg and by extending at the hip joint. In addition, the thigh of the non-support leg aids in force production by flexing at the hip joint. Upon landing, the force is absorbed by flexion at the hips and the supporting knee. The arms participate vigorously in force production as they move up and down in a bilateral manner. Due to the vigorous action and precarious balance of performers at this stage, the number of hops generally ranges between two and four.

Stage 3 The thigh of the non-support leg is in a vertical position with the knee flexed at 90 degrees or less. Performers exhibit greater body lean forward than in stages one or two, with the result that the hips are farther in front of the support leg upon take-off. This forward lean of the trunk results in greater distance in relation to the height of the hop.

The thigh of the non-support leg remains near the vertical (frontal) plane, but knee flexion may vary as the body is projected and received by the supporting leg. The arms are used in force production, moving bilaterally upward during the force production phase.

Stage 4 The knee of the non-support leg is flexed at 90 degrees or less, but the entire leg swings back and forth like a pendulum as it aids in force production. The arms are carried close to the sides of the body, with elbow flexion at 90 degrees. As the non-support leg increases its force production, that of the arms seems to diminish.

NAME_____DATE_____CLASS_____

Find the CORRECT HEADING or title and write it in the blank. Then place one of these on following lines: NOT DISCUSSED, NOT ACCURATE, TOO BROAD, TOO NARROW, (a heading may be used more than once.)

Paragraph 1

French Guiana is, for the most part, wild and primitive. It is a backward country unspoiled by modern trends and the least explored of the three Guianas. At one time it was known for its infamous Devil's Island Penal Colony, but that has been deactivated. The country is rich in minerals and hardwood timbers. In Cayenna visitors can see carts drawn by water buffalo and trees covered with exquisit wild orchids.

1. Facts about French Guiana_____
2. Devil's Island Penal Colony_____
3. Minerals and Hardwood in French Guiana_____
4. The Most Explored of the Three Guianas _____
5. The Races in French Guiana_____

Paragraph 2

The eight-hour day is universal both in fields and factories of the Hawaiian sugar plantations. Employees have leisure time to work in their gardens. They make full use of recreational facilities at the clubs, athletic fields, and swimming pools. Movies are shown nightly at the plantation theatres. Practically all of the homes have radios and a high proportion have washing machines and other modern equipment. One out of every three employees owns a car.

1. The Eight-Hour Work Day _____
2. The Hawaiian Sugar Industry _____
3. Schools on Hawaiian Sugar Plantations_____
4. Living Conditions in Hawaiian Plantation Communities_____
5. Educational Level of Plantation Workers_____

Paragraph 3

Maize formed the staff of life of the early North American Indians. One of its remarkable forms was zuni corn which, in order to resist drought, was planted 18 inches deep. The Pueblo Indians used corn in many of their ceremonies such as the snake dance, and in their corn planting and harvesting they observed certain rituals handed down from their forefathers. The prayers for rain and the methods of cultivation of the maize in separate clumps argue for a very ancient use of this valuable plant.

1. Cultivation and Use of Corn by Early North American Indians_____
2. Early American Food Plants_____
3. The Use of Corn in Indian Ceremonials_____
4. The Preservation of Corn by Drying_____
5. The Planting of Zuni Corn_____

Paragraph 4

We are accustomed to the thought that geographic conditions have profoundly influenced wars in the past. Washington's army escaped the "Generals January and February" in his advance against Russia. The Valley of the Shenandoan and the Blue Ridge had important bearing on our Civil War. The great distance of the Philippines from home and the tropic character of its climate were important factors in our war with the Filipinos.

1. Our Changing Climate_____
2. General January and February_____
3. Escape in a Fog_____
4. Geographic Influences on Wars of the Past_____
5. The Importance of Geographic Factors_____

Paragraph 1

The pygmy of Central Africa is traditionally a forest dweller. He considers the forest his own and resents any intrusion by outsiders. The men in the tribe are hunters. The women do the fishing, gather wild vegetables from the forest and do all the cooking. The children carry the water. There is no pottery in their culture but they do make bows and arrows and spears, do some basketry, and build thatched huts. They eat both raw and cooked meat and make fire by twirling sticks. They wear a short apron of bark or a girdle of leaves, they may paint their faces with a dark fluid. The women paint their bodies in geometric designs, and the paint is never washed off. In fact, pygmies never wash.

Choose the best statement: CIRCLE THE LETTER

a. It was the writer's main purpose to explain the small size and poor condition of the pygmies.
b. It was the writer's main purpose to explain and excuse the unfriendliness of the pygmies toward intruders.
c. It was the writer's main purpose to describe the pygmies and their ways of life.
d. It was the writer's main purpose to gain sympathy for the pygmy in his struggle to exist.

Paragraph 2

The history of English is full of examples of words that have changed their psychological meaning and have gradually come to have meanings that are sinister. One of the modern examples of this is the word "graft". The verb "to graft" meant at first merely to work. The specialized meaning of digging seems to be implied in the Anglo-Saxon form from which the modern word is descended. Early colloquial English used the word in such expressions as "Where are you grafting?" meaning "Where do you live or work?" From this perfectly respectable meaning the word has gradually gone down hill. The work of which the grafter is accused today is not of the sort originally expressed; it is rather of a sort which fills the name "grafter" with opprobrium.

1. The function of this paragraph is

a. to numerate details
b. to tell a story; to narrate
c. to explain
d. to compare

1._____

2. Choose the best meaning for sinister

a. open to question
b. different from the original
c. evil
d. whimsical

2._____

3. Choose the best meaning for opprobrium

a. respect
b. disgrace
c. amusement
d. doubt

3._____

4. Choose the best central thought

a. The word "graft" is one of many words in English that have changed meaning and gone down hill.
b. A study of the history of words shows that over long periods of time most words take on sinister meanings.
c. Today the work of the "grafter" covers the word with shame but there was a time when the meaning was entirely respectable.
d. The history of English is full of examples of words that have come to us from the Anglo-Saxon.

4._____

Consistent or Inconsistent C or I

1. The child is older than her parent. _____
2. Many are more than a few. _____
3. An angry man makes his friends comfortable. _____
4. A likely result is quite probable. _____
5. A moving object is a stationary item. _____
6. Malicious motives usually will produce kind acts. _____
7. A day will follow the longest night. _____
8. If we continue to add, we will enhance the total. _____
9. One cannot consider his family among his friends. _____
10. It is difficult for a nation to have both complete freedom and absolute regulation. _____

Mark the similar meaning with S the opposite meaning with O

Example: 1. The event will take place in February.

____ a. We were thinking of expanding.

____ b. We are expecting a winter happening.

____ c. If should occur in Brazil.

____ d. It is planned for the end of the year.

1. The speech set them laughing.
____a. Everything was in order
____b. The audience howled in glee.
____c. The remarks deepened the gloom.
____d. No item could be considered.
2. She made a flashy appearance.
____a. It was a striking entrance.
____b. This is a true spirit.
____c. She has a big heart.
____d. She looked quite plain.
3. The victory was not gained fairly.
____a. No man is an island.
____b. We do not plan to lose.
d. It was a dishonest effort.